Product name:	ATP7B Rabbit Polyclonal Antibody
Cat number:	ABN07345
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human ATP7B. AA range:161-210
Reactivity:	Human,Mouse,Rat
Applications:	IHC 1:100-1:300,ICC/IF 1:200-1:1000,ELISA 1:5000-1:10000
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

This gene is a member of the P-type cation transport ATPase family and encodes a protein with several membrane-spanning domains, an ATPase consensus sequence, a hinge domain, a phosphorylation site, and at least 2 putative copper-binding sites. This protein functions as a monomer, exporting copper out of the cells, such as the efflux of hepatic copper into the bile. Alternate transcriptional splice variants, encoding different isoforms with distinct cellular localizations, have been characterized. Mutations in this gene have been associated with Wilson disease (WD). [provided by RefSeq, Jul 2008],catalytic activity:ATP + H(2)O + Cu(2+)(In) = ADP + phosphate + Cu(2+)(Out).,disease:Defects in ATP7B are the cause of Wilson disease (WD) [MIM:277900]. WD is an autosomal recessive disorder of copper metabolism in which copper cannot be incorporated into ceruloplasmin in liver, and cannot be excreted from the liver into the bile. Copper accumulates in the liver and subsequently in the brain and kidney. The disease is characterized by neurologic manifestations and signs of cirrhosis.,function:Involved in the export of copper out of the cells, such as the efflux of hepatic copper into the bile.,online information:Wilson's disease website,PTM:Isoform 1 may be proteolytically cleaved at the N-terminus to produce the WND/140 kDa form.,similarity:Belongs to the cation transport ATPase (P-type) family.,similarity:Belongs to the cation transport ATPase (P-type) family. Type IB subfamily.,similarity:Contains 6 HMA domains.,subcellular location:Predominantly found in the trans-Golgi network (TGN). Not redistributed to the plasma membrane in response to elevated copper levels.,subunit:Monomer. Interacts with COMMD1/MURR1.,tissue specificity:Most abundant in liver and kidney and also found in brain. Isoform 2 is expressed in brain but not in liver. The cleaved form WND/140 kDa is found in liver cell lines and other tissues.,